Case Reports

Primary Malignant Fibrous Histiocytoma of the Right Ventricle and Main Pulmonary Trunk

with a Review of the Literature

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We report the case of a 76-year-old man with a malignant fibrous histiocytoma of the right ventricle and main pulmonary trunk, diagnosed through echocardiography and catheterization. Extensive resection of the right ventricular outflow tract, pulmonary valve apparatus, and main pulmonary trunk was performed, and the defect was repaired with a valveless ventriculo-pulmonary Dacron graft. The patient recovered uneventfully, and was doing well 18 months after surgery.

To our knowledge, this is only the 15th case of primary malignant fibrous histiocytoma of the heart that has been documented in the literature since histologic criteria and cases were published in 1977-78, and the 2nd such case of a primary tumor that has arisen in a right cardiac chamber. The case is presented in detail, along with a review of the literature since 1978. ***(Texas Heart Institute Journal 1989;16:296-304)***

Primary sarcoma of the right ventricle and the main pulmonary trunk is rare. We report such a case, diagnosed in a 76-year-old man. Upon resection and pathologic examination, the lesion proved to be a malignant fibrous histiocytoma (MFH)—an entity that, although commonly found in the soft tissue of elderly men,[1] seldom presents as a primary cardiac tumor. Fourteen other cases of primary cardiac MFH have been documented in the literature since the 1977-78 publication of MFH cases and diagnostic criteria,[1-4] and in only one of these did the primary tumor arise in a right cardiac chamber. We present the details of our case, along with a review of the literature.

Key words: *Histiocytoma, malignant fibrous; heart neoplasms; neoplasm recurrence, local; heart ventricle, right; sarcoma; tumor of the main pulmonary trunk; cardiopulmonary bypass; ventriculo-pulmonary bypass graft*

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Case Report

In January of 1987, a 76-year-old man was referred to our hospital for recurrent right ventricular failure. In addition to having diabetes mellitus and mild hypertension, he had a 1-month history of progressive dyspnea, lower-limb edema, and hepatodynia without fever. There was no history of syncope, angina, or weight loss. Upon treatment with diuretic and inotropic drugs, the symptoms disappeared and the patient was discharged from the hospital. Because of an unexplained recurrence of right-sided cardiac failure, however, he was readmitted on an emergency basis in March of 1987.

2nd Admission

Upon clinical examination at readmission, the patient was found to be dyspneic (New York Heart Association functional class III), with a pulse of 100 beats per minute and a blood pressure of 135/90 mmHg. His jugular veins were distended, and he had painful hepatomegaly and extensive edema of the lower limbs. His lungs were clear. Auscultation revealed a normally situated apical impulse. The 1st heart sound was normal, but the pulmonic component of the 2nd heart sound was inaudible. A grade 3/6 systolic ejection murmur was heard at the 2nd and 3rd left intercostal spaces. The chest radiograph showed a cardiothoracic ratio of 60% and a prominent pulmonary knob. The electrocardiogram showed sinus rhythm, a frontal plane QRS axis of +120°, an "S1 - Q3" feature in the precordial leads, an

incomplete right bundle-branch block, right atrial and ventricular overload, and right ventricular hypertrophy. Blood gas analysis indicated hypoxemia (72 mmHg) and hypocapnia (30 mmHg) at rest, which initially suggested chronic thromboembolic disease. The results of phlebo-iliocavography were normal. A lung scan disclosed hypoperfused areas in the apex of the right lower pulmonary lobe and in the anterior segment of the right upper lobe. M-mode and 2-dimensional echocardiography showed markedly enlarged, hypocontractile right cardiac cavities and exaggerated motion of the tricuspid valve leaflets. A short-axis view indicated an obstructive mass filling the right ventricular outflow tract (RVOT) and the main pulmonary trunk, and extending into the left and right pulmonary arteries (Fig. 1). At right cardiac catheterization, right ventricular pressure was 100/25 mmHg, and pulmonary artery pressure was 20/5 mmHg, with a gradient of 80 mmHg across the pulmonary valve. The cardiac index was 1.8 L/min/m^2. Right cardiac angiography was not performed.

We diagnosed a tumor of the main pulmonary trunk and the RVOT. Because of the patient's critical hemodynamic state, we began exploratory surgery. After performing a sternotomy and pericardiotomy, we could see the tumor on the myocardium of the RVOT and the main pulmonary trunk. The mass was marked by invasive white lobulated areas. The right cardiac cavities were enlarged. We instituted cardiopulmonary bypass with moderate (27 °C) hypothermia , and protected the myocardium by means of cold cardioplegia and topical pericardial cooling. After aortic cross-clamping, we incised the long axis of the RVOT vertically through the pulmonary annulus, and extended this incision to the main pulmonary trunk. This exposed a large, glistening, gelatinous, white-gray multilobular mass, which protruded, with non-adhesive extension, into the right and left main pulmonary arteries (Fig. 2A). The mass also extended into the right ventricular cavity, nearly occluding the pulmonary valve, and damaging it beyond effective repair. There was no clear line of division between the tumor and the main pulmonary trunk; extensive resection became necessary, and involved the entire pulmonary trunk, the pulmonary valve, and the anterior aspect of the RVOT. The resected tumor measured 10 x 2 x 3 cm and weighed 75 g.

We repaired the defect with a valveless, 30-mm-diameter ventriculo-pulmonary woven-Dacron tube graft, cutting its proximal segment obliquely to cover the resected anterior aspect of the RVOT and securing it with continuous mattress sutures. The distal segment of the graft was anastomosed end-to-end to the bifurcation of the main pulmonary trunk (Fig. 2B). After 65 minutes of aortic cross-clamping, sinus rhythm was induced, the heart was rewarmed, and cardiopulmonary bypass was discontinued. The remainder of the procedure was performed routinely.

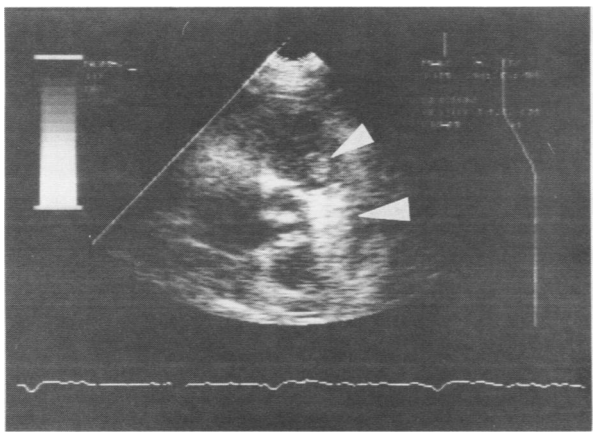

Fig. 1 *A short-axis echocardiographic view of the great vessels at the base of the heart. The tumor mass fills the right ventricular outflow tract and the main pulmonary trunk (arrows).*

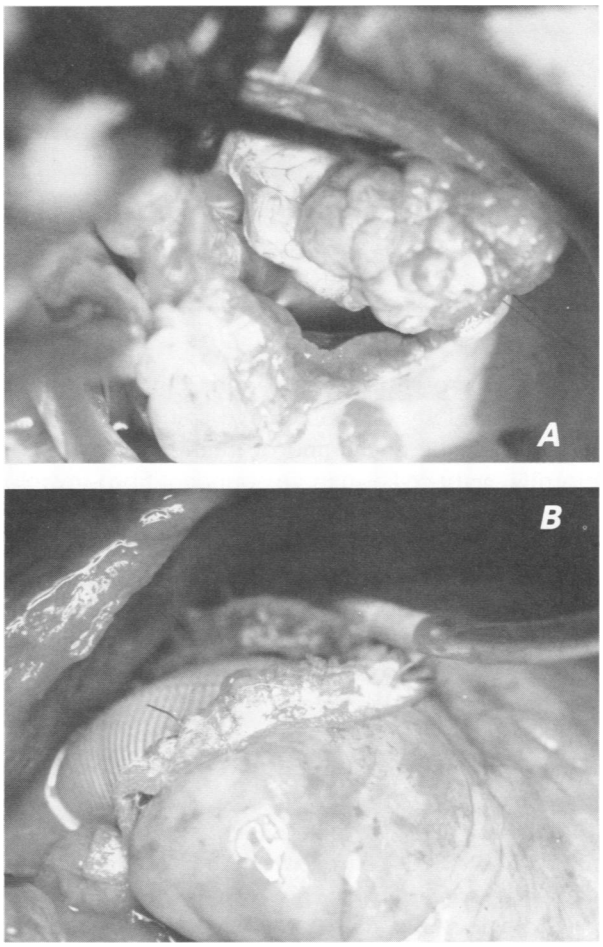

Fig. 2 *Intraoperative photographs show **A**) the lobulated, gelatinous white-gray tumor in the resected right ventricular outflow tract, and **B**) the ventriculo-pulmonary bypass with a valveless Dacron tube graft.*

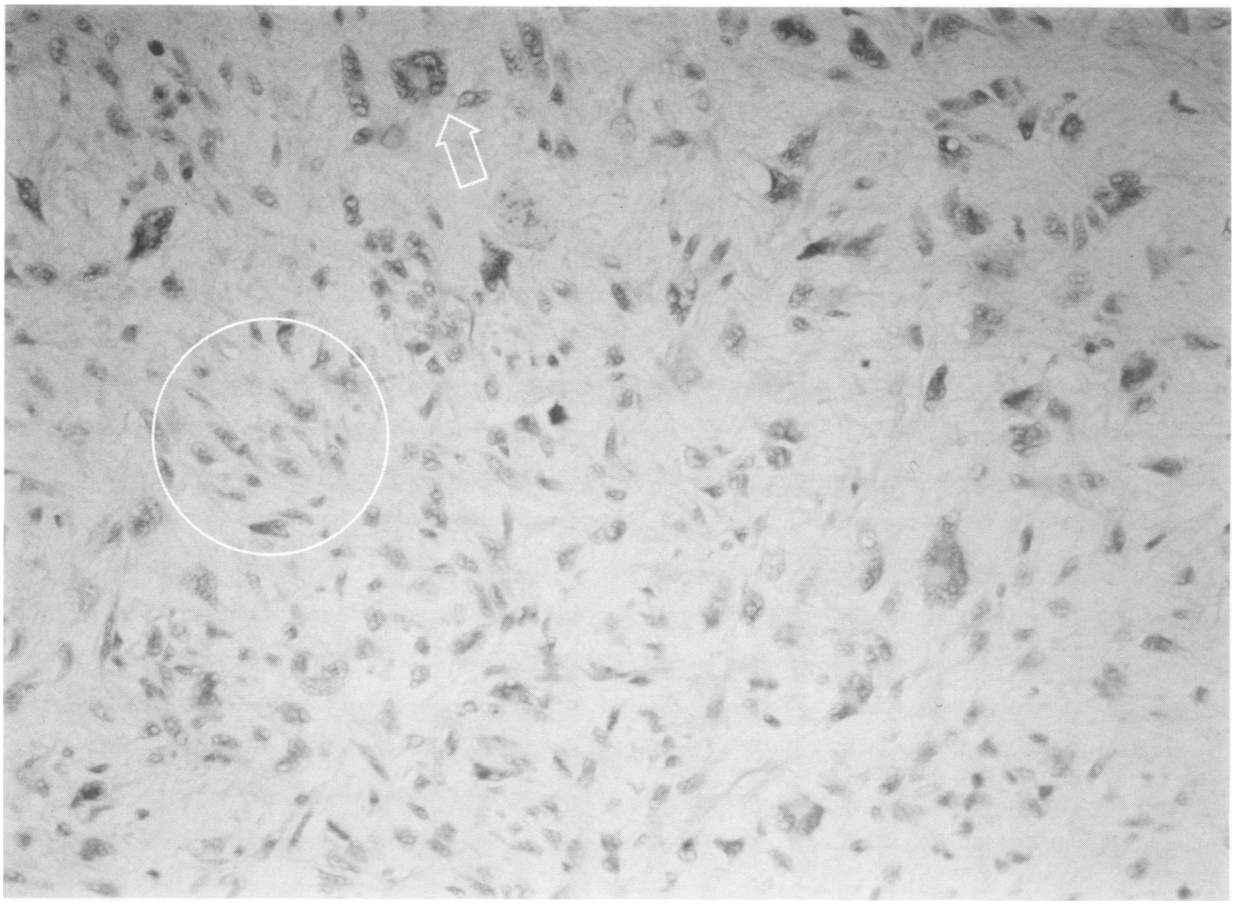

Fig. 3 *Photomicrograph of the resected malignant fibrous histiocytoma. The tumor contains cells resembling both fibroblasts and histiocytes. While there are pleomorphic giant cells (arrow) and spindle cells (circle), note the absence of inflammatory cells. (H + E, orig. x250)*

Upon microscopic examination (Fig.3), the tumor consisted chiefly of proliferative spindle cells with hyperchromatic nuclei and of multinucleated pleomorphic giant cells, all in an abundant fibro-myxoid stroma. The giant cells had nuclei of irregular shape, and often of large size; nucleoli were prominent. Mitotic figures were occasionally encountered, and mitotic nuclear abnormalities confirmed the malignancy of the tumor. There were no rhabdomyosarcomatous or leiomyosarcomatous differentiating features. The diagnosis rendered was malignant fibrous histiocytoma, myxoid variant. The margins of the resected right ventricular outflow tract were positive for the neoplasm.

The patient's postoperative course was uneventful, and he was discharged from the hospital 12 days after surgery. Because of his advanced age and poor physiologic status, he received no chemotherapy or radiotherapy. Postoperative echocardiography showed satisfactory right ventricular ejection and graft patency. At follow-up 18 months later, the patient had returned to a nearly normal lifestyle with no recurrence of major symptoms.

Discussion

Although MFH, thought to arise from primitive mesenchymal cells, is a relatively common malignant tumor of the connective tissue in adults, its occurrence in the heart is extremely unusual. The differentiation of MFH is quite recent:[1-4] since 1978, only 14 cardiac cases of primary MFH have been reported[5-19] (Table I).

Morphology

Gross Appearance. Macroscopically, MFH is a sessile or pedunculated tumor, multilobulated or polypoid; in color it is gray, white, yellow, or red, or intermediate shades of those colors. The growth has a smooth, glistening, gelatinous surface; its point of attachment is upon occasion calcified, as reported by Mori[11] and Laya.[18] Cross-section typically shows areas of necrosis, intratumoral hemorrhage, or both. In the cases we review below, the tumor commonly measures 5 cm or more in diameter.

Microscopic Appearance. Under microscopic examination, the lesion classically presents a storiform

pattern of fibroblast-like spindle cells and histiocyte-like round or polygonal cells, accompanied by pleomorphic giant cells and inflammatory cells.[18] Pathologic diagnosis requires that there be no specific features of differentiation other than collagen production and phagocytosis.[18,20]

Myxoid Variant. The 4 morphologic subgroups of MFH that have been described are myxoid, storiform-pleomorphic, giant cell, and inflammatory.[2,18] In their 1977 study[3] that proposes a myxoid subgroup of MFH for its potential usefulness in establishing prognosis, Weiss and Enzinger indicate that the degree of myxoid change appears to be inversely related to the rate of metastasis: i.e., the more myxoid, the lower the probability of metastasis. Although this observation was made without specific application to primary cardiac MFH, it possibly helps account for the absence of metastasis in our patient. The same study, however, reports a strong tendency among predominately myxoid MFH tumors to lose their myxoid character by becoming progressively cellular with each new recurrence—so the probability of metastasis would increase in later manifestations of the primary neoplasm.

Review of the Literature

In our review of the literature since 1978, we include only first publication of cases, and exclude broad reviews of institutional experience that lack specific dates of occurrence or histologic documentation.[21,22] We also exclude a very early case of fibrous histiocytoma reported by Rose,[23,24] and a later case resolved by heart transplantation,[25] because they lack criteria for a diagnosis of malignancy. The only other published case[26] of which we are aware is one cited in Kern,[13] about which we have been unable to obtain direct information: this is unfortunate, for discrete primary tumors are reported to have been found both in the left atrium and in the right ventricle.

A review of the 15 cases in Table I shows that the patients' mean age was 44.1 years (range, 21 to 77 years). Eight (53%) of the patients were women. Thirteen (86%) of the primary tumors were encountered in the left atrium:[5-12,14-19] 10 of these arose from the posterior or posterolateral wall;[7-12,14,15,18,19] 1 arose from the interatrial septum;[5,6] and in the remaining 2 cases, the tumor grew around the annulus of a mitral valve prosthesis.[16,17] Pericardial extension was observed in 4 left atrial tumors;[7,11,15,17] extension to the right or left pulmonary veins was seen in 2 such tumors;[12,14] and obstruction of the 4 pulmonary veins in 1 tumor.[7] Both the right-sided MFH tumor reported by Kern and colleagues[13] and the one we report here are exceptional in that they arose not from a wall or septum, but in Kern's patient from the pulmonary valve apparatus and in our patient from the RVOT with pulmonary extension: both tumors, in fact, extended across the pulmonary valve toward the main pulmonary trunk. Eight of the 15 patients (53%) had probable or confirmed metastasis.

Symptoms Arising from Primary Condition. The most common symptoms on presentation were those associated with the primary condition, which usually manifested itself as congestive heart failure: dyspnea, orthopnea, asthenia, syncope, productive cough, and edema, alone or in some combination. In patients with left-heart tumors, these symptoms were on occasion aggravated by arrhythmia[7,11,12] or by mitral regurgitation.[17] However, some patients first presented with symptoms arising from conditions secondary to the tumor.

Symptoms Arising from Secondary Conditions. As would be expected, most secondary symptoms could be traced to distal embolism of tumor or thrombus, to metastasis, or to metastatic embolism. Embolism to the central nervous system manifested itself as transient cerebral ischemic attack,[16] hemiparesis,[7,16,18] aphasia,[16] and dysarthria.[7] Gastrointestinal symptoms—epigastralgia,[9,15] nausea,[15] and hematemesis[15]—arose in response to metastasis, which in Minamiji's patient[15] presented as distal tumor with intussusception of the jejunum.

Pulmonary symptoms arising from apparent or confirmed thromboembolism were found in both patients whose primary tumors originated on the right side of the heart. The patient described by Kern and associates[13] was first treated for *Staphylococcus aureus* pneumonia, after presenting with severe cough, fever, and left pleuritic pain. Subsequently he underwent a lobectomy for persistent pulmonic cavitation, but surgical discovery thereupon of unilateral pulmonary thromboembolism and infarct led only to a passing suspicion of extrapulmonic neoplasm. Actual diagnosis was not made until autopsy, 4 years later. This case is extraordinary in that the primary tumor was extremely slow in its growth. More commonly, MFH grows so rapidly that secondary symptoms do not appear in advance of primary symptoms, and in any event do not obscure the primary condition for long. Our own patient, on lung scan, showed hypoperfusion of the right upper and lower pulmonary lobes, a condition possibly caused by distal embolism; but symptoms of right ventricular failure arising from the primary condition were far more prominent.

Upon occasion polyarthritis occurs as a 1st manifestation of occult malignant disease, with no evidence of distal embolism or metastasis. Berkelbach and coworkers[14] report a patient whose initial symptoms were those of paraneoplastic nonspecific polyarthritis, manifested months before onset of congestive heart failure. A 21-year-old man presented with fatigue, lethargy, and a 5-month history of pain and swelling in his knees, wrists, and ankles. Observation

TABLE I. Primary Cardiac Fibrous Histiocytoma: Review of the Literature

Author	Year of Publication	Age/Sex	Primary Tumor		Complaints on Presentation	Diagnostic Techniques	Operative Treatment	Other Therapy	Extensions, Metastases	Number of Recurrences	Outcome (mo)
			Origin	Size or Weight							
Shah[5] Gabelman[6]	1978 1979	37/F	LA (IAS)	2x2x5 cm	Dyspnea; weight gain; edema; productive cough	Echo; R-L cath; angio	Resection	Radio; late chemo		4	Death (33)
Hamada[7]	1980	33/F	LA (posterior wall)	5 cm	Dyspnea; arrhythmia; hemiparesis; dysarthria; orthopnea; hemoptysis; edema	Echo; R-L cath; angio	Partial resection		Extension to pericardium; apparent systemic embolism		Death (3)
Morimoto[8]	1980	36/M	LA (posterior wall)	child's fist				Radio	Metastases to both lungs and right pleura		Death (48)
Terashima[9]	1983	27/F	LA (postero-lateral wall)	man's fist	Asthenia; epigastralgia; dyspnea; edema; pericardial syndrome	Autopsy		Radio; chemo	Extension to IAS and RA, with metastases to left adrenal and left cervical regions, jejunum		Death (27)
Concha[10]	1983	30/M	LA (posterior wall)	large	CHF	Echo	Resection	Chemo; late radio	Extension to atrial septum	1	Death (21)
Mori[11]	1983	61/F	LA (posterior wall)	50 g	Dyspnea; arrhythmia; syncope; asthenia	Echo; CT scan; R Cath; angio	Resection		*	*	Death (6)
Eckstein[12]	1984	27/F	LA (posterior wall)	10x10 cm	Asthenia; low-grade fever; dyspnea; pleural effusion; arrhythmia	Echo; CT scan; cath; angio	Resection	Chemo	Extension to R pulmonary vein; metastasis to brain at 22 mo (excised)	2	Well (24)

Kern[13]	1985	77/M	RV (base of PV)	4x4x3 cm	Cough; fever; left pleuritic pain	Autopsy			Metastatic thromboembolism to lung, with hemorrhagic infarct		Death (ca. 50)
Berkelbach[14]	1985	21/M	LA (posterior wall)	6x4x4 cm	Arthralgia due to poly-arthritis; asthenia	Echo	Partial resection		Extension into left pulmonary vein	1	Death (6)
Minamiji[15]	1986	45/F	LA (postero-lateral wall)		Epigastralgia; nausea; hemate-mesis; intussus-ception of jejunum; CHF	Echo; R-Cath	Partial resection	Chemo	Extension into pericardium; metastasis to jejunum, right adrenal gland	1	Death (4)
Holtzman[16]	1986	68/F	LA (in assoc. with MV prosthesis)	2x2x5 cm	CHF; weight loss; low-grade fever; TIAs; R hemi-paresis; aphasia	Autopsy			Extension into myocardium; small artery embolism		Death (ca. 12)
Lee[17]	1987	65/M	LA (in assoc. with MV prosthesis)		Hemiparesis; mitral regur-gitation	Autopsy	MVR		Extension to pericardium; metastasis to brain		Death (ca. 5)
Laya[18]	1987	28/F	LA (posterior wall)	3 masses totaling 57 g	Dyspnea; orthopnea; asthenia; productive cough	Echo	Resection	Chemo	Metastases to liver, brain, skeleton	1	Death (14)
Binon[19]	1988	30/M	LA (posterior wall)	4x5 cm	Weight loss; asthenia	Echo	Partial resection		Extension to pulmonary vein		Death (periop)
Glock	1989	76/M	RV (RVOT)	10x2x3 cm	CHF; hepato-dynia; lower-limb edema	Echo; cath	Resection		Extension into pulmonary trunk, destroying PV		Well (18)

Angio = angiocardiography; Cath = cardiac catheterization; CHF = symptoms associated with congestive heart failure; Chemo = chemotherapy; CT = computed tomographic; Echo = echocardiography; IAS = interatrial septum; IVS = interventricular septum; LA = left atrium; mo = months after presentation; MV = mitral valve; MVR = mitral valve replacement; periop = perioperative; PV = pulmonary valve; R = right; Radio = radiotherapy; R = right; R-L = right and left; RV = right ventricle; RVOT = right ventricular outflow tract; TIA = transient ischemic attack (cerebral)

* A 2nd tumor, with pericardial extension, arose subsequently on the anterior wall of the right ventricle, but was considered an extension of the original tumor on the posterior wall of the left atrium, rather than a metastasis.

of an enlarged cardiac silhouette on chest radiography led to echocardiographic diagnosis of a cardiac mass. Even at recurrence of the primary tumor, there was no evidence of skeletal metastasis.

Diagnostic Methods. The interval between onset of symptoms and diagnosis was variable, ranging from a few months to 4 years or longer.[7,13] Diagnosis of a cardiac mass (if not specifically of MFH) was made during life in at least 10 patients,[5-7,10-12,14,15,18,19] including ours, and after death in 4 patients.[9,13,16,17] In 7 patients, cardiac auscultation was of initial help in suggesting a tumor: in instances of left atrial tumor, a diastolic mitral murmur[5-7,18] or a "tumor plop"[7] was sometimes heard. An inflammatory syndrome was present in 7 patients,[5-7,11,12,14,15,17] and Terashima's group[9] reported thrombocytopenia.

Echocardiographic findings were positive (most clearly so in 2-dimensional mode) for a cardiac mass in 10 cases[5,6,10-12,14,15,18,19] including our own, but are reported to have given false-negative results in at least 2[13,16] of the 4 cases that were not diagnosed until postmortem examination.

Before resection of the initial tumor, 6 of the 10 patients whose echocardiographic findings had been positive underwent confirmatory or definitive diagnostic procedures. Confirmation was by catheterization, with or without angiocardiography, but 1 patient also underwent computed tomographic (CT) scanning before the 1st operative procedure: in the case reported by Mori and associates,[11] CT scanning with the use of contrast medium enabled better definition of the size and location of the initial tumor.

Two other patients (of the 6 who had undergone catheterization) were subjected to CT scanning, but only upon tumor recurrence. In Eckstein's patient,[12] CT scanning helped reveal a new point of tumor attachment, and obstruction of the right pulmonary vein. In Minamiji's patient,[15] CT scanning of the recurrent tumor was followed by scintigraphy, which showed massive uptake by the tumor of gallium-67 and thallium-201 tracers (except, the authors speculate, in areas of necrosis and hemorrhage).

In the remaining patients with positive echocardiographic results,[10,14,18,19] the first operative procedure was undertaken after diagnosis by echocardiography alone. While confirmatory procedures often helped to define a tumor's size and exact location, positive findings of the initial echocardiograms were in all 10 cases supported by subsequent investigative and therapeutic procedures. Clinicians, nevertheless, should give serious consideration to angiocardiography in patients older than 40 years, in order to evaluate the coronary arteries before surgery.

Therapy. Although surgical resection of malignant fibrous histiocytoma is the treatment of choice, it still appears to be palliative at best, due largely to local extension of the primary neoplasm. In our patient, we found the borders of the resected RVOT to be positive for MFH, even after we had performed broad resection and reconstruction.

Of the 11 patients who had surgical treatment, 1 died perioperatively.[19] Four patients required emergency operation: a young man whose hemodynamic status was critical;[10] an older woman with severe pulmonary congestion;[11] a man with intractable pulmonary edema and deteriorating consciousness;[17] and a young woman, recently delivered of a child by caesarian section, who was suffering progressive cardiac decompensation.[18]

Treatment of local recurrences varied. Two patients with poor prognoses received no further therapy;[14,15] 1 patient underwent 3 reoperations for recurrent tumors;[5,6] 1 underwent chemotherapy alone;[18] 1 had radiotherapy alone, after an adjuvant course of chemotherapy (Adriamycin, DTIC-Dome, and vincristine) had failed to prevent recurrence;[10] and 1 underwent chemotherapy and surgery for successive tumors.[12]

The patient who underwent both chemotherapy and surgery was a 27-year-old woman whose case was reported by Eckstein and colleagues.[12] They used chemotherapy alone to treat a 1st recurrence at 3 months after initial surgery. This successfully reduced the tissue mass of the neoplasm to 2 cm, yet the tumor recurred again at 14 months. On 2nd recurrence, broad resection was undertaken (interatrial septum, portion of left atrial wall, entire right lung, and paratracheal and paraesophageal lymph nodes), followed by GORE-TEX reconstruction of the atrial septum and wall. Eckstein's patient was alive and well at more than 2 years after initial presentation, and the authors recommended that adjuvant chemotherapy (the combined CY-VA-DIC regimen) follow every surgical excision of a cardiac soft tissue sarcoma. Although we believe that chemotherapy, radiotherapy—or both in combination—can be helpful, the interactions and results of these therapies are difficult to predict. In our patient's case, we decided against adjuvant chemotherapy because of his advanced age and debilitated condition.

Metastatic tumors were surgically excised when they appeared as single, resectable entities.[12,15]

Prognosis. Because of the likelihood of metastasis or local recurrence, patients with MFH of the heart have a poor prognosis. At least 7 of the 15 reported cases involved metastasis. The rate of local recurrence was even higher: 6 patients[5,6,10,12,14,15,18] of the 9 who survived initial surgery had at least 1 recurrence during the 1st postoperative year. Shah and Gabelman's patient[5,6] had 4 recurrent tumors over 33 months, despite aggressive triple therapy.

For the 14 patients who did not die perioperatively, mean survival time after initial presentation was 19 months. Five patients survived for 2 years or

longer: 1 for at least 24 months, with chemotherapy followed by radical resection and reconstruction;[12] 1 for 27 months, with chemotherapy and radiotherapy;[9] 1 for 33 months, with multiple resections and the administration of both chemotherapy and radiotherapy;[5,6] 1 for 48 months, with radiotherapy alone;[8] and 1 for approximately 50 months, with no treatment specific to the primary neoplasm, which went undetected and was unusually slow in its growth.[13]

Addendum

The patient was hospitalized in October of 1989 for superior-vena-cava-obstruction syndrome. Computed tomographic scanning, upper-limb phlebography, and magnetic resonance imaging showed extensive local regrowth of the tumor, with thromboembolism to the right pulmonary artery and superior vena cava. Reoperation was contraindicated, and drug therapy was initiated for symptomatic relief. The patient died at the end of November 1989 of cardiac failure and metastasis to the axillary and cervical lymph nodes. His survival time, without adjuvant chemotherapy or radiotherapy, was approximately 35 months after initial presentation and 33 months after resection and reconstruction.

References

1. Weiss SW, Enzinger FM. Malignant fibrous histiocytoma: an analysis of 200 cases. Cancer 1978;41:2250-66.
2. Enzinger FM. Recent developments in the classification of soft tissue sarcomas. In: Management of primary bone and soft tissue sarcomas. Chicago: Year Book Medical Publishers, 1977: 219-234.
3. Weiss SW, Enzinger FM. Myxoid variant of malignant fibrous histiocytoma of the heart. Cancer 1977;39:1672-85.
4. McAllister HA, Fenoglio JJ. Fibrosarcoma and malignant fibrous histiocytoma. In: McAllister HA, Fenoglio JJ, eds. Tumors of the cardiovascular system: Fascicle 15, 2nd series, Atlas of tumor pathology. Washington: Armed Forces Institute of Pathology, 1978:95-9.
5. Shah AA, Churg A, Sbarbaro JA, Sheppard JM, Lamberti J. Malignant fibrous histiocytoma of the heart presenting as an atrial myxoma. Cancer 1978;42:2466-71.
6. Gabelman C, Al-Sadir J, Lamberti J, et al. Surgical treatment of recurrent primary malignant tumor of the left atrium. J Thorac Cardiovasc Surg 1979;77:914-21.
7. Hamada N, Matsuzaki M, Kusukawa R, et al. Malignant fibrous histiocytoma of the heart. Jpn Circ J 1980;44:361-8.
8. Morimoto K, Yumoto T, Matsui K, Yoshida H, Hashimoto K, Irisawa T. Primary malignant fibrous histiocytoma of the heart [in Japanese with English abstract]. Jpn J Cancer Clin 1980;26: 81-6.
9. Terashima K, Aoyama K, Nihei K, et al. Malignant fibrous histiocytoma of the heart. Cancer 1983;52:1919-26.
10. Concha M, Pasalodos J, Rubio FL. Surgical treatment of primary malignant fibrous histiocytoma of the left atrium. Int J Cardiol 1983;4:463-6.
11. Mori K, Itoh H, Kanaya H, et al. Malignant fibrous histiocytoma of the heart. Jpn Circ J 1983;47:188-93.
12. Eckstein R, Gössner W, Rienmüller R. Primary malignant fibrous histiocytoma of the left atrium: surgical and chemotherapeutic management. Br Heart J 1984;52:354-7.
13. Kern SE, Cowen ME, Abrams GD. Malignant fibrous histiocytoma of the heart presenting as unilateral pulmonary thromboembolism and infarct. Hum Pathol 1985;16:1279-81.
14. Berkelbach van der Sprenkel JW, Timmermans AJM, Elbers HRJ, van Herpen G, Dinant HJ. Polyarthritis as the presenting symptom of a malignant fibrous histiocytoma of the heart. Arthritis Rheum 1985;28:944-7.
15. Minamiji K, Yamasaki T, Nakao M, Furumoto M, Yoshida Y. 201-Tl and 67-Ga uptake in malignant fibrous histiocytoma of the heart. Chest 1986;89:303-6.
16. Holtzman E, Schiby G, Segal P, Priel I. Malignant fibrous histiocytoma complicating mitral valve replacement. J Am Coll Cardiol 1986;7:956-60.
17. Lee J, Cheung KL, Wang R, Mok CK, Khin MA. Malignant fibrous histiocytoma of the left atrium. J Thorac Cardiovasc Surg 1987;94:450-2.
18. Laya MB, Mailliard JA, Bewtra C, Levin HS. Malignant fibrous histiocytoma of the heart: a case report and review of the literature. Cancer 1987;59:1026-31.
19. Binon JP. Histiocytomes fibreus malins cardiaques. Clermont Ferrand. Universite de Clermont I, 1988:104 p.
20. Weis LM, Warhof MJ. Ultrastructural differentiation between adult pleomorphic rhabdomosarcoma, pleomorphic liposarcoma and pleomorphic malignant fibrous histiocytoma. Human Pathol 1984;15:1025-33.
21. Reece IJ, Cooley DA, Frazier OH, Hallman GL, Powers PL, Montero CG. Cardiac tumors: clinical spectrum and prognosis of lesions other than classical benign myxoma in 20 patients. J Thorac Cardiovasc Surg 1984;88:439-46.
22. Dein JR, Frist WH, Stinson EB, et al. Primary cardiac neoplasms: early and late results of surgical treatment in 42 patients. J Thorac Cardiovasc Surg 1987;93:502-11.
23. Rose AG. Fibrous histiocytoma of the heart [letter to the editor]. Arch Pathol Lab Med 1978;102:389.
24. Fabian JT, Rose AG. Tumours of the heart: a study of 89 cases. S Afr Med J 1982;61:71-7.
25. Jamieson SW, Gaudiani VA, Reitz BA, Oyer PE, Stinson EB, Shumway NE. Operative treatment of an unresectable tumor of the left ventricle. J Thorac Cardiovasc Surg 1981;81:797-9.
26. Omura K, Magara T, Iwa T, et al. Emergent surgery for malignant fibrous histiocytoma of the heart. Rinsho Kyobu Geka 1983;3:356.

Editorial Comment

Fibrous histiocytomas may often be diagnosed by endomyocardial biopsy, since approximately 50% have an intracavitary component. However, histological features play a disappointingly small role in predicting the biological behavior of these neoplasms. The presence of certain atypical histological features, including necrosis, marked cellularity, and mitotic activity, does not correlate well with metastatic potential. Indeed, metastasis may occur from fibrous histiocytomas that lack all of the foregoing features.

Because of the lack of certainty of malignant behavior of these neoplasms by histological criteria alone, and the poor prognosis in those patients with metastatic fibrous histiocytoma, aggressive surgical resection is indicated whenever feasible. Of the 14

patients with cardiac fibrous histiocytomas in the AFIP series,[1] 9 had multiple sites of involvement within the heart; however, only 7 of the 14 patients had extracardial spread of the neoplasm at autopsy (5 with distant metastases; 2 with direct spread to adjacent structures). Therefore, in those patients without evidence of extracardial spread in whom tumor resection is not possible, cardiac transplantation may be considered as a therapeutic option even in the presence of histological criteria for malignancy.

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Reference

1. McAllister HA, Fenoglio JJ. Fibrosarcoma and malignant fibrous histiocytoma. In: McAllister HA, Fenoglio JJ, eds. Tumors of the cardiovascular system: Fascicle 15, 2nd series, Atlas of tumor pathology. Washington: Armed Forces Institute of Pathology, 1978:95-9.